

Rheumatic Heart Disease

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Objectives

- ❖ Epidemiology of the disease
- ❖ Pathogenesis of rheumatic heart disease
- ❖ Clinical features: cardiac & non-cardiac manifestations
- ❖ Laboratory studies for RHD
- ❖ Principles of management

Acute rheumatic fever

- Autoimmune consequence of infection with Group A streptococcal infection
- Results in a generalised inflammatory response affecting brains, joints, skin, subcutaneous tissues and the heart.
- The clinical presentation can be vague and difficult to diagnose.
- Currently, the modified Duckett-Jones criteria form the basis of the diagnosis of the condition.

Rheumatic heart disease

- Rheumatic diseases are defined by the constellation of results of the physical examination, autoimmune marker and other serologic tests, tissue pathology, and imaging.
- Recognition of clinical patterns remains essential for diagnosis because there is no single diagnostic test and results may be positive in the absence of disease.

Rheumatic heart disease

- ❖ Rheumatic Heart Disease is the permanent heart valve damage resulting from one or more attacks of ARF.
- ❖ 40-60% of patients with ARF will go on to developing RHD.
- ❖ The commonest valves affecting are the mitral and aortic,....

Causes

- ❖ Rheumatic fever is thought to result from an inflammatory autoimmune response.
- ❖ Rheumatic fever only develops in children and adolescents following group A beta-hemolytic streptococcal pharyngitis, and only streptococcal infections of the pharynx initiate or reactivate rheumatic fever.
- ❖ Strong correlation between progression to rheumatic heart disease and human leukocyte antigen (HLA)-DR class II alleles and the inflammatory protein-encoding genes *MBL2* and *TNFA*.

Pathophysiology

- ❖ **Group A streptococci elaborate the cytolytic toxins streptolysins S and O.**
- ❖ **Of these, streptolysin O induces persistently high antibody titers that provide a useful marker of group A streptococcal infection and its nonsuppurative complications**

Pathophysiology

- ❖ In 0.3-3% of cases, infection leads to rheumatic fever several weeks after the sore throat has resolved.
- ❖ Patients remain infected for weeks after symptomatic resolution of pharyngitis and may serve as a reservoir for infecting others.
- ❖ Rheumatic fever develops in some children and adolescents following pharyngitis with group A beta-hemolytic Streptococcus

Pathophysiology

- ❖ The organisms attach to the epithelial cells of the upper respiratory tract and produce a battery of enzymes allowing them to damage and invade human tissues.
- ❖ After an incubation period of 2-4 days, the invading organisms elicit an acute inflammatory response with 3-5 days of sore throat, fever, malaise, headache, and an elevated leukocyte count

Pathophysiology

- ❖ Acute rheumatic heart disease often produces a pancarditis characterized by endocarditis, myocarditis, and pericarditis.
- ❖ Endocarditis is manifested as valve insufficiency.
- ❖ The mitral valve is most commonly and severely affected (65-70% of patients), and the aortic valve is second in frequency (25%).
- ❖ The tricuspid valve is deformed in only 10% of patients and is almost always associated with mitral and aortic lesions.
- ❖ The pulmonary valve is rarely affected.
- ❖ Pericarditis, when present, rarely affects cardiac function or results in constrictive pericarditis

Epidemiology

- ❖ At this time, rheumatic fever is uncommon among children in the western countries but common in the arabian countries.
- ❖ Incidence of rheumatic fever and rheumatic heart disease has decreased in the United States and other industrialized countries.
- ❖ Decreased incidence of rheumatic fever has been attributed to the introduction of penicillin or a change in the virulence of the *Streptococcus*.

❖ Mortality/Morbidity

- ❖ The disease is more severe in females than in males. Insufficiency from acute rheumatic valve disease resolves in 60-80% of patients who adhere to antibiotic prophylaxis

❖ Race

- ❖ Native Hawaiian and Maori (both of Polynesian descent) have a higher incidence of rheumatic fever (13.4 per 100,000 hospitalized children per year), even with antibiotic prophylaxis of streptococcal pharyngitis.

❖ Sex

- ❖ Rheumatic fever occurs in equal numbers in males and females, but the prognosis is worse for females than for males.

❖ Age

- ❖ Rheumatic fever is principally a disease of childhood, with a median age of 10 years, although it also occurs in adults (20% of cases).

CLINICAL PRESENTATIONS

HISTORY

- ❖ A diagnosis of rheumatic heart disease is made after confirming antecedent rheumatic fever.
- ❖ The Jones criteria require the presence of 2 major or 1 major and 2 minor criteria for the diagnosis of rheumatic fever.
- ❖ The major diagnostic criteria include carditis, polyarthrititis, chorea, subcutaneous nodules, and erythema marginatum.

A. Major criteria:

- Carditis.
- Polyarthriti
- Sydenham's chorea.
- Erythema marginatum.
- Subcutaneous nodules.

B. Minor criteria:

- Previous history of rheumatic fever.
- Arthralgia.
- Fever.
- Lab tests indicative of inflammation : ESR (erythrocyte sedimentation rate), CRP (C-Reactive protein), leukocytosis.
- ECG changes.

PHYSICAL FINDINGS

- ❖ **Physical findings in a patient with rheumatic heart disease include cardiac and noncardiac manifestations of acute rheumatic fever.**
- ❖ **Some patients develop cardiac manifestations of chronic rheumatic heart disease**

CARDIAC MANIFESTATIONS

- ❖ Pancarditis is the most serious and second most common complication of rheumatic fever (50%).
- ❖ Patients may complain of dyspnea, mild-to-moderate chest discomfort, pleuritic chest pain, edema, cough, or orthopnea.
- ❖ Upon physical examination, carditis is most commonly detected by a new murmur and tachycardia out of proportion to fever.
- ❖ New or changing murmurs are considered necessary for a diagnosis of rheumatic valvulitis.

- The following murmurs are most commonly observed during acute rheumatic fever:
 - Apical pansystolic murmur is a high-pitched, blowing-quality murmur of mitral regurgitation that radiates to the left axilla.
 - Apical diastolic murmur (also known as a Carey-Coombs murmur) is heard with active carditis and accompanies severe mitral stenosis..
 - Basal diastolic murmur is an early diastolic murmur of aortic regurgitation and is high-pitched, blowing, decrescendo, and heard best along the right upper and mid-left sternal border after deep expiration while the patient is leaning forward.

- Congestive heart failure
 - Heart failure may develop secondary to severe valve insufficiency or myocarditis.
 - Tachypnea, orthopnea, jugular venous distention, rales, hepatomegaly, a gallop rhythm, edema, and swelling of the peripheral extremities.
- Pericarditis
 - A pericardial friction rub indicates that pericarditis is present.
 - Increased cardiac dullness to percussion and muffled heart sounds are consistent with pericardial effusion.
 - A paradoxical pulse (and accentuated fall in systolic blood pressure with inspiration) with decreased systemic pressure and perfusion

NON CARDIAC MANIFESTATIONS

- Common noncardiac manifestations of acute rheumatic fever include:
 - ❖ polyarthrititis, chorea, erythema marginatum, and subcutaneous nodules.
 - ❖ Other clinical, noncardiac manifestations include abdominal pain, arthralgias, epistaxis, fever, and rheumatic pneumonia.
- Polyarthrititis is the most common symptom and is frequently the earliest manifestation of acute rheumatic fever (70-75%).
- The arthritis reaches maximum severity in 12-24 hours, persists for 2-6 days (rarely more than 3 wk) at each site, and rapidly responds to aspirin.
- Sydenham chorea occurs in 10-30% of patients with rheumatic fever.

- ❖ A long latency period (1-6 mo) between streptococcal pharyngitis and the onset of chorea is observed.
- ❖ Patients with chorea often do not demonstrate other Jones criteria. It is also known as rheumatic chorea, Sydenham chorea, chorea minor, and St Vitus dance.
- ❖ Daily handwriting samples can be used as an indicator of progression or resolution of disease.
- ❖ Complete resolution of the symptoms typically occurs with improvement in 1-2 weeks and full recovery in 2-3 months.

- ❖ Erythema marginatum, also known as erythema annulare, is a characteristic rash that occurs in 5-13% of patients with acute rheumatic fever.



- ❖ Subcutaneous nodules are currently an infrequent manifestation of rheumatic fever.
- ❖ When present, the nodules appear over the extensor surfaces of the elbows, knees, ankles, knuckles, and on the scalp and spinous processes of the lumbar and thoracic vertebrae where they are attached to the tendon sheath.
- ❖ These nodules are strongly associated with severe rheumatic carditis, and, in the absence of carditis, the diagnosis of subcutaneous nodules should be questioned.

LABORATORY STUDIES

LABORATORY STUDIES

❖ Throat culture

- ❖ Throat culture findings for group A beta hemolytic *Streptococcus* are usually negative by the time symptoms of rheumatic fever or rheumatic heart disease appear.

❖ Rapid antigen detection test

- ❖ This test allows rapid detection of group A streptococcal antigen

❖ Antistreptococcal antibodies

- ❖ The clinical features of rheumatic fever begin at the time antistreptococcal antibody levels are at their peak.
- ❖ The most common extracellular antistreptococcal antibodies tested include
 - ❖ antistreptolysin O (ASO),
 - ❖ antideoxyribonuclease (DNAse) B,
 - ❖ antihyaluronidase,
 - ❖ antistreptokinase,
 - ❖ antistreptococcal esterase
 - ❖ anti-DNA.

- ❖ Acute phase reactants

- ❖ The C-reactive protein and erythrocyte sedimentation rate are elevated in rheumatic fever due to the inflammatory nature of the disease. Both tests have a high sensitivity but low specificity for rheumatic fever..

- ❖ Heart reactive antibodies

- ❖ Tropomyosin is elevated in acute rheumatic fever.

- ❖ Rapid detection test for D8/17

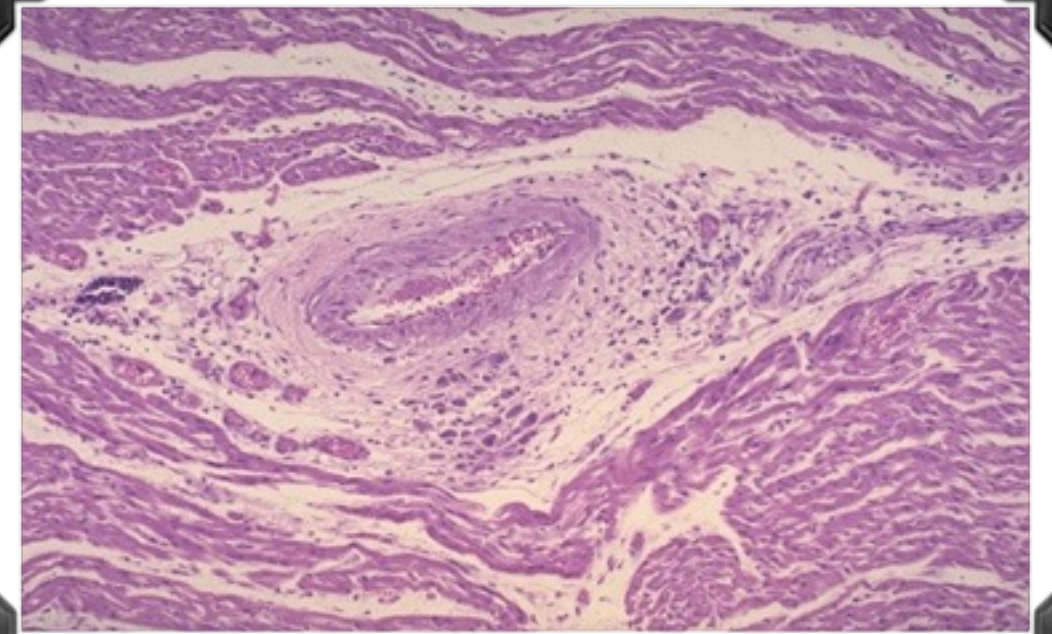
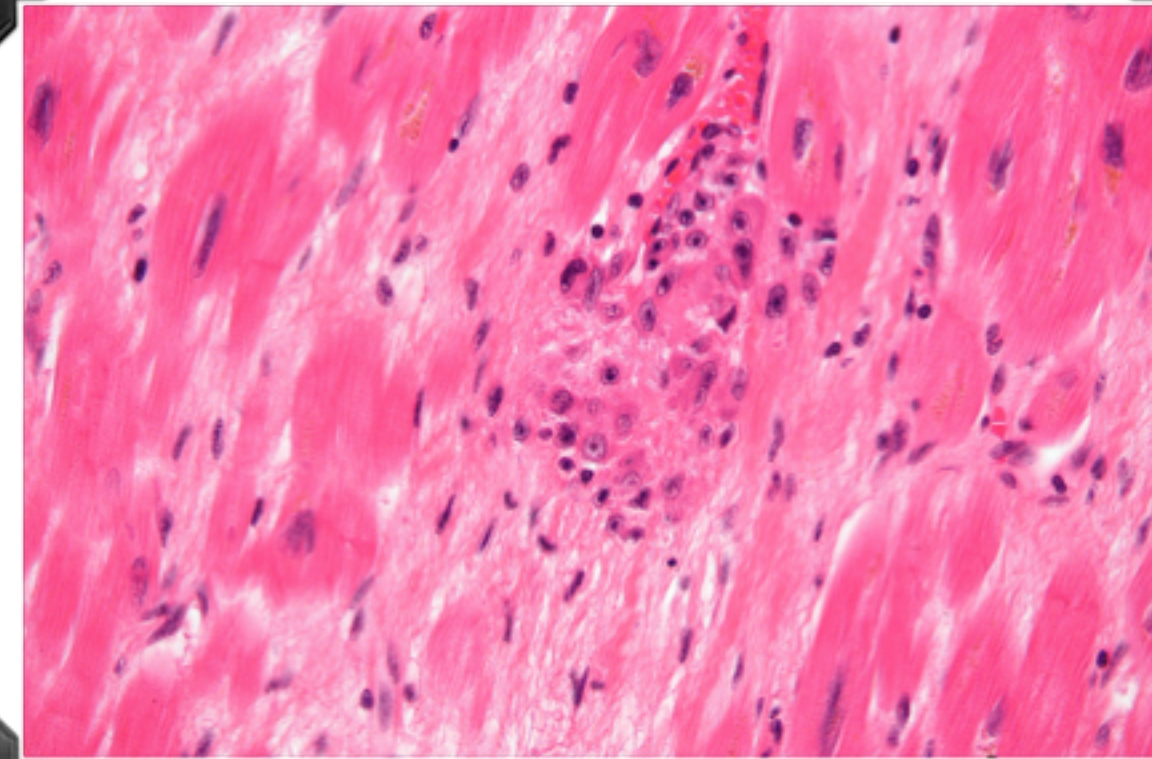
- ❖ This immunofluorescence technique for identifying the B cell marker D8/17 is positive in 90% of patients with rheumatic fever

Investigation

- ❖ ECG
- ❖ Chest roentgenography
- ❖ Echocardiography

PATHOLOGY

- ❖ Pathologic examination of the insufficient valves may reveal verrucous lesions at the line of closure.
- ❖ Aschoff bodies (perivascular foci of eosinophilic collagen surrounded by lymphocytes, plasma cells, and macrophages) are found in the pericardium, perivascular regions of the myocardium, and endocardium.



Acute phase:

acute rheumatic pancarditis (inflammation of endocardium, myocardium and pericardium)

1. Myocarditis.

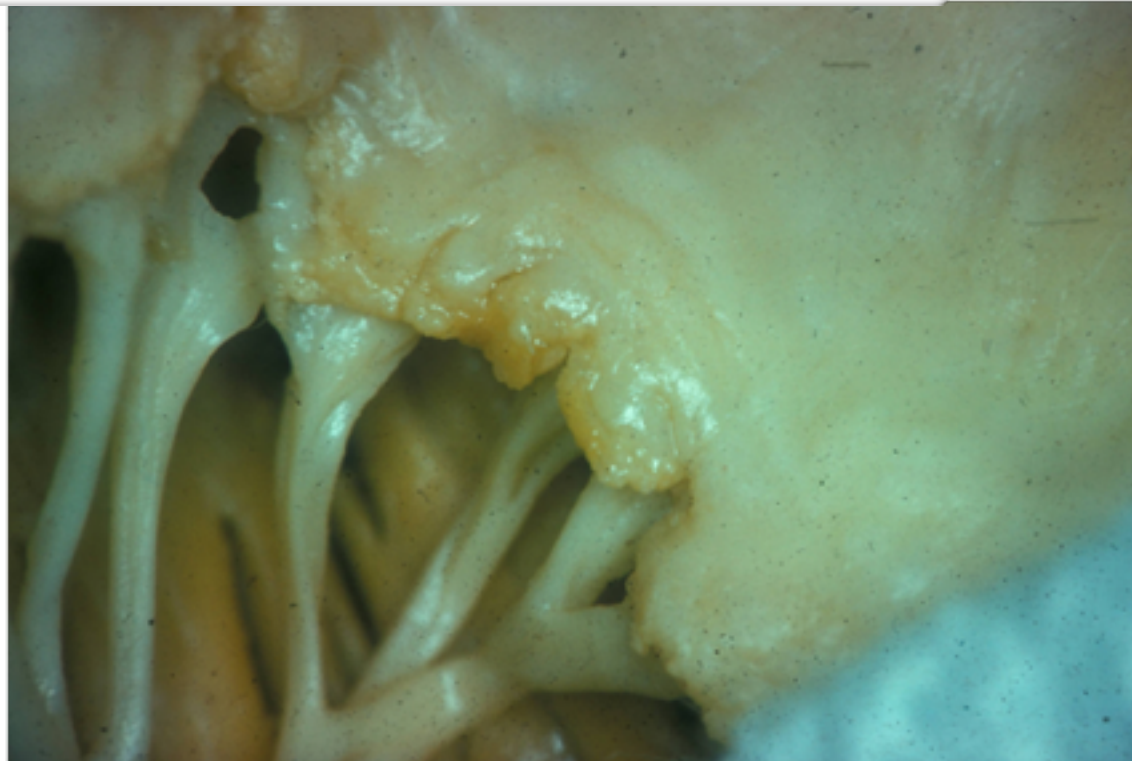
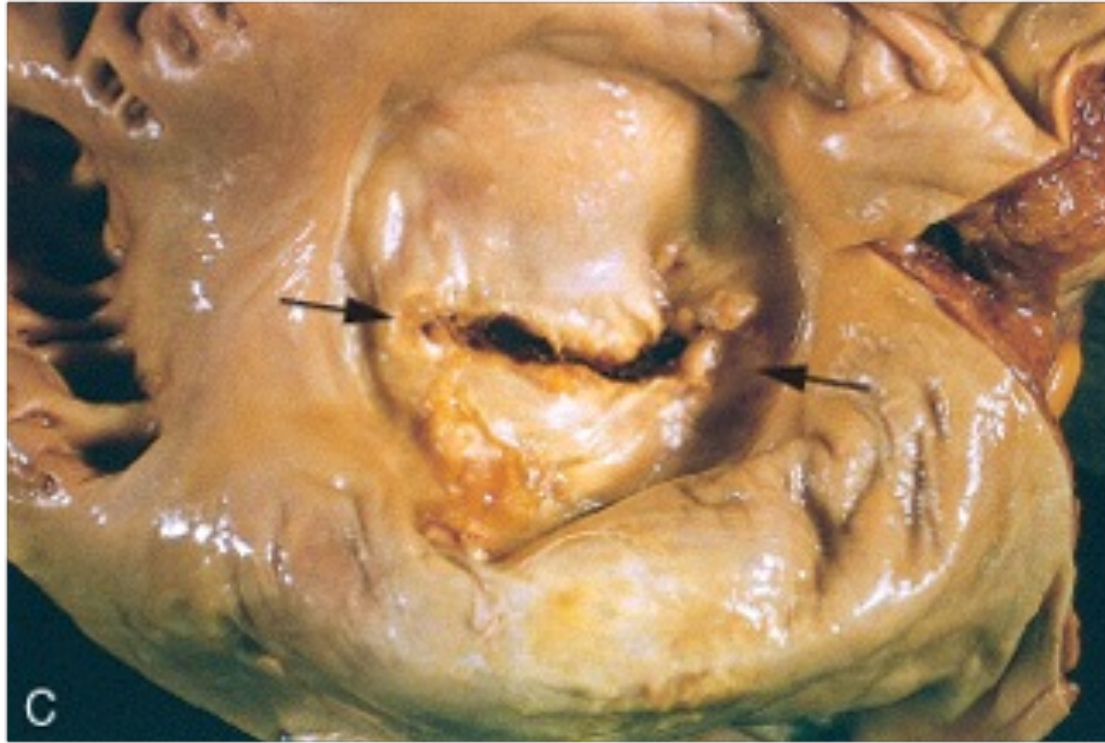
2. Pericarditis: "bread and butter", due to fibrinous inflammation

3. Endocarditis: edema, inflammation and fibrin deposits on valve leaflets (vegetations) along lines of closure. Mostly mitral and aortic valves. Aschoff nodules are uncommon in the valves.

Chronic phase:

Acute changes may resolve completely or progress to scarring and development of chronic valvular deformities many years after the acute disease.

RHEUMATIC MS





MANAGEMENT

- ❖ therapy is directed toward eliminating the group A streptococcal pharyngitis (if still present), suppressing inflammation from the autoimmune response, and providing supportive treatment for congestive heart failure.
- ❖ Following the resolution of the acute episode, subsequent therapy is directed towards preventing recurrent rheumatic heart disease in children and monitoring for the complications and sequelae of chronic rheumatic heart disease in adults.

Prevention of rheumatic fever in patients with group A beta hemolytic streptococci (GABHS) pharyngitis

- ❖ For patients with GABHS pharyngitis,
- ❖ Oral (PO) penicillin V remains the drug of choice for treatment of GABHS pharyngitis, but ampicillin and amoxicillin are equally effective.
- ❖ When PO penicillin is not feasible or dependable, a single dose of intramuscular benzathine penicillin G or benzathine/procaine penicillin combination is therapeutic.
- ❖ For patients who are allergic to penicillin, administer erythromycin or a first-generation cephalosporin. Other options include clarithromycin for 10 days, azithromycin for 5 days, or a narrow-spectrum (first-generation) cephalosporin for 10 days. As many as 15% of patients who are allergic to penicillin are also allergic to cephalosporins.

- ❖ Aspirin in anti-inflammatory doses effectively reduces all manifestations of the disease except chorea, and the response is typically dramatic.
- ❖ If rapid improvement is not observed after 24-36 hours of therapy, question the diagnosis of rheumatic fever.

- ❖ Include digoxin and diuretics, afterload reduction, supplemental oxygen, bed rest, and sodium and fluid restriction as additional treatment for patients with acute rheumatic fever and heart failure.
- ❖ The diuretics most commonly used in conjunction with digoxin for children with heart failure include furosemide and spironolactone.
- ❖ Initiate digoxin only after checking electrolytes and correcting hypokalemia.
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- ❖ Afterload reduction (ie, using ACE inhibitor captopril) may be effective in improving cardiac output, particularly in the presence of mitral and aortic insufficiency.
- ❖ Start these agents judiciously. Use a small, initial test dose (some patients have an abnormally large response to these agents), and administer only after correcting hypovolemia.
- ❖ When heart failure persists or progresses during an episode of acute rheumatic fever in spite of aggressive medical therapy, surgery is indicated and may be life-saving for severe mitral and/or aortic insufficiency.

- ❖ The duration of antibiotic prophylaxis is controversial.
- ❖ Continue antibiotic prophylaxis indefinitely for patients at high risk (eg, health care workers, teachers, daycare workers) for recurrent GABHS infection.
- ❖ Patients with rheumatic fever with carditis and valve disease should receive antibiotics for at least 10 years or until age 40 years.

Surgical care

- ❖ In patients with critical stenosis, mitral valvulotomy, percutaneous balloon valvuloplasty, or mitral valve replacement may be indicated.
- ❖ Due to high rates of recurrent symptoms after annuloplasty or other repair procedures, valve replacement appears to be the preferred surgical option





Thank you